

CONGENITAL HEART BLOCK :

REPORT OF A CASE

BY

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The comparative rarity of congenital heart block as recorded in medical journals warrants the addition of another case to this collection. Up to 1938 approximately 55 cases have been described.

There is no doubt that one of the reasons for not discovering these cases early in childhood is that signs and symptoms are usually absent or only minimal, and that the heart rate is inclined to be more rapid than in cases with acquired complete heart block (Campbell and Suzman, 1934). It is only when some other medical condition brings such a person to a physician that the heart block may be discovered. Sometimes this person may be examined by several competent doctors before the defect is accidentally stumbled upon. This is illustrated by the presentation of the following case.

A white, female child, aged 3 years, was brought to the hospital in February 1940 with the complaint of a purulent discharge from the right ear of one year's duration. This had started four months after an uneventful convalescence from measles, when she had been treated by a physician who made no comment about her heart. Two previous attacks of acute bronchitis also requiring the attention of a physician had caused no comment relative to her heart. Otherwise she had always been a normal-looking, well-nourished, and healthy, active youngster, without any cardiac signs or symptoms. She was a full-term baby, delivered normally, and experienced no difficulties in her babyhood. A clinical diagnosis of right acute mastoiditis was made. There were no other findings at this time. The intern made a note that he heard no murmurs over the praecordium but did not record the pulse or cardiac rate. The nurse's chart showed a pulse rate of 80. She had an operation on the same day.

During the entire time of operation the anaesthetist noted the pulse rate as being 56, and brought it to the surgeon's attention. Her condition despite this rate was good. The oto-laryngologist commented on the possibility of the low rate being due to an intradural abscess, and after completing a simple mastoidectomy asked for a neurological consultation.

Two days later the neurologist reported that he found no neurological signs, and suggested that a paediatrician should look for some congenital anomaly because of the bradycardia.

It was then that a pædiatrician (J. S. U.) first saw her and noted "an extensive bradycardia and a low musical murmur, systolic in time, heard all over the præcordium, but loudest at the apex and fourth left intercostal space near the sternum." This was thought to be a murmur that could be accounted for most logically on the basis of a patent interventricular septum being present, inasmuch as after it had been heard it had always been present and had never varied in intensity, quality, or location.

An electrocardiogram revealed an auricular rate of 120, a ventricular rate of 70, complete heart block, a tendency to left axis deviation, and a diphasic T wave in lead IV F (Fig. 1).

Fluoroscopic and radiographic examination of the heart revealed enlargement of both the right and left ventricles and left auricle (Fig. 2). The lung

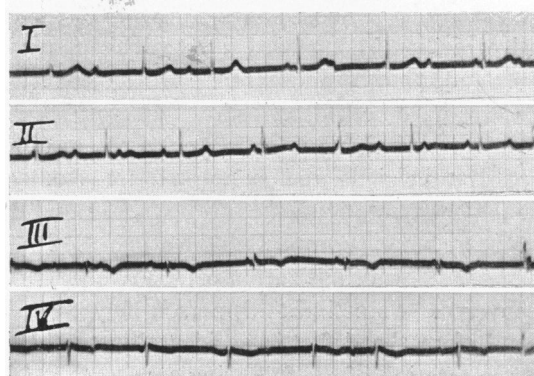


FIG. 1.—Electrocardiogram showing complete heart block (A, 120; V, 70).

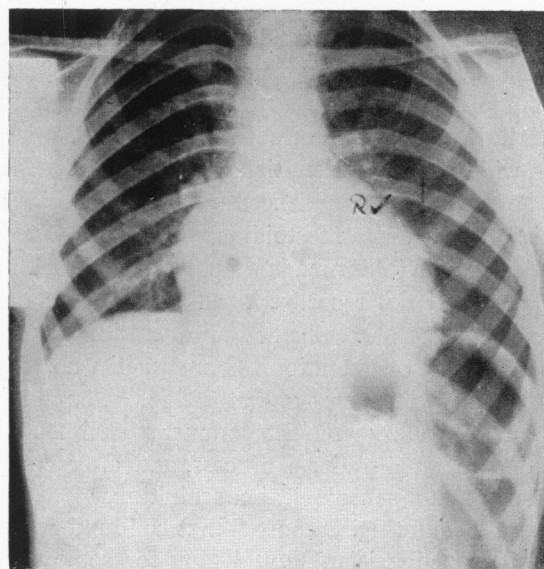


FIG. 2.—Radiogram of heart showing enlargement of the right and left ventricles.

fields were clear. Barium in the œsophagus in the right (I) oblique view showed an indentation of the œsophagus from a moderately enlarged left auricle.

Other laboratory findings were as follows. There was no abnormality of the urine except for a faint trace of albumin. Complete blood count: haemoglobin, 78 per cent; red blood corpuscles, 4,560,000; white blood corpuscles, 9050, with a normal differential count.

The rectal temperature on admission was 99.6° F. and varied from 100 to 99 until the patient was discharged.

The nurse's chart recorded a pulse average of from 56 to 52 during the patient's stay in hospital. The respirations averaged from 24 to 28 per minute.

The patient responded well to the operation and was discharged after two weeks, returning to the out-patient department for mastoid surgical dressings and follow-up in the pædiatric clinic. Two months later another electrocardiogram during one of her visits to the out-patient department again showed complete heart block.

Her convalescence from the right mastoidectomy was uneventful.

DISCUSSION

Complete heart block represents an inability of the stimulating impulses originating in the sino-auricular node to pass through the auriculo-ventricular node and the main bundle of His, so that the auricles beat at their own rate while the ventricles contract at the rate governed by the auriculo-ventricular node that has taken over the function of pacemaker, which is usually 30 to 50 beats per minute, or if conduction from the latter is impaired also, then at their own ventricular rate, which is usually below 40 beats per minute. This heart block may be either congenital, which is rare, or acquired, which is more usual.

Yater (1929) reviewed 30 cases of congenital heart block reported up to that time, and established certain criteria that would permit one to classify a case as being congenital. These were the following five points:

1. Electrocardiographic evidence of the block existing.
2. A slow pulse that had been found present at an early age and had continued to be present.
3. The absence of any history suggestive of an infection that might have produced the block, as diphtheria, congenital or early acquired syphilis, rheumatic fever, or chorea.
4. A history of any one of the following: syncopal attacks, fainting spells (explained on the basis of the Adams-Stokes syndrome), vertigo, headaches, nausea and vomiting after unusual exertion, convulsions, dyspnoea, and/or cyanosis. (These findings may or may not be present and are not absolutely requisite for the criteria, but if present add further evidence.)
5. The presence of a congenital heart lesion, namely, patent interventricular septum (the latter at times being subject to an acquired endo-myocarditis, quite infrequent in this instance as far as the septum itself is concerned).

While theoretically it might be possible to explain the heart block on a pre-natal myocarditis or syphilis involving the bundle of His or a developmental

defect affecting the bundle of His, these are most unlikely to occur for practical considerations.

This case fits the criteria of Yater (1929).

The ætiology of congenital heart block is organic and is based on the pathological presence of a patent interventricular septum. Such septal defects, both large and small, are a common deformity and are found relatively frequently. Why they occur clinically without the presence of heart block so often, is odd, but has been explained correctly as follows : The usual sight of the interventricular defect is anterior to the pars membranacea while the A-V bundle lies behind it (Leech, 1930). Inasmuch as the ventricular complexes in the electrocardiogram of complete heart block are normal, it follows that the lesion responsible for the block must be in the course of the main bundle of His, above the bifurcation close to the septum (Lampard, 1928).

The reason for the almost constant preservation of the muscular connection between the auricles and ventricles probably lies in the fact that the special bundle appears in the fifth week of foetal life, whereas the membranous separation between auricles and ventricles take form between seven and ten weeks. The bundle is preserved between the posterior endocardial cushion and the posterior portion of the annular fibrosis (Yater, Leaman, and Cornell, 1934; Moll, 1912; and Tandler, 1913).

It is only when the A-V node or main bundle of His is caught in the congenital anomalous development of the septum, or is caught in the excessive formation of fibrous tissue of the membranous portion of the septum interfering with the continuity of the bundle, that heart block occurs (Aitken, 1932).

The prognosis in general is guarded. A few cases have reached mature adult life. It must be remembered that a patent interventricular septum is associated often with other congenital cardiac anomalies, the most frequent being the tetralogy of Fallot. These anomalies are overshadowed by the dangers of an engrafted endocarditis. If the concomitant anomalies are small and do not limit the functional capacity of the heart permitting the subject to survive early life, there is no reason to consider the prognosis unfavourable, barring an unlooked-for bacterial endocarditis.

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